Rev. Inst. Med. trop. S. Paulo
51(1):53-55, January-February, 2009
doi: 10.1590/S0036-46652009000100010

CASE REPORT

ENDOBRONCHIAL LEIOMYOMA: AN UNUSUAL NON-DEFINING NEOPLASM IN A PATIENT WITH AIDS

Humberto METTA(1), Marcelo CORTI(1), Liliana REDINI(1), Roberto DURE(1), Ana M. CAMPITELLI(1) & Marina NARBAITZ(2)

SUMMARY

Smooth muscle neoplasms are more frequent in human immunodeficiency infected children than in HIV seropositive adults. Endobronchial leiomyoma is a rare benign tumor in HIV infected adult patients. Epstein-Barr virus (EBV) has been implicated in the pathogenesis of these tumors. Here we describe an adult patient with HIV infection with atelectasis of the left upper pulmonary lobe as the first clinical expression of an intrabronchial leiomyoma. In this case, we can not show the association with EBV. Our report suggests that smooth muscle tumors as leiomyoma should be included in the differential diagnosis of endobronchial masses in AIDS patients.

KEYWORDS: Endobronchial leiomyoma; Atelectasis; AIDS; HIV.

INTRODUCTION

Epstein-Barr virus-associated smooth muscle tumors (EBV-SMT) include leiomyomas and leiomyosarcomas which are the second most prevalent malignancy in children with AIDS[7]. These tumors are not very common in adults and can be located in the soft tissue[3] and in the gastrointestinal tract[10] and less frequently in the liver[14], the spleen[1], the adrenal gland, the urinary tract including the kidneys and the gallbladder[16] and the central nervous system (meninges and spinal neural canal)[12]. EBV-SMT in adults with AIDS are extremely rare, with only few cases involving the respiratory tract reported in the literature[6].

We present a case of an adult HIV/AIDS patient who developed an endobronchial leiomyoma without evidence of an association with EBV.

CASE REPORT

A 36 year-old HIV seropositive man with diagnosis of AIDS on account of history of cryptococcal meningitis and *Pneumocystis jiroveci* pneumonia, presented with fever (39 °C), progressive dyspnea, productive cough and weigh loss (approximately 5 k weight during the last month). The chest radiograph revealed atelectasis of the left upper lobe plus a left pleural effusion. A computed tomography showed the same findings (Fig. 1). The CD4 T cell count was 9 cells/μL (3%). Flexible fiberoptic bronchoscopic examination disclosed a left endobronchial lobulated tumor that obstructed the ostium of the left upper lobe, as the cause of the atelectasis (Fig. 2). It was completely removed and atelectasis was resolved. Histopathologic examination revealed the smooth muscle origin of this tumor by the routine microscopy. The diagnosis was confirmed by means of a positive immunostaining for smooth muscle actin. Histologic features were mesenchymal tumor characterized by the fusocellular proliferation of spindle cells with cigar-shaped nuclei, eosinophilic fibrillar cytoplasms

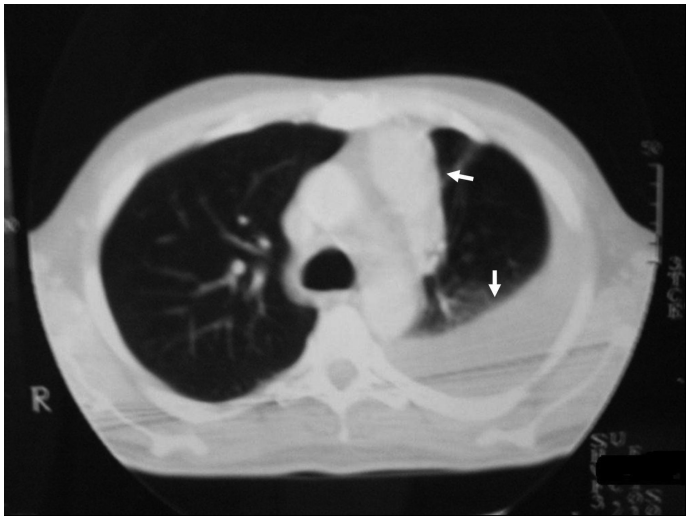

Fig. 1 - A thorax computed tomography showing the left pleural effusion and the atelectasis of the left upper lobe (arrows). It is shown the different intensity in the opacity of atelectasis in comparison with the pleural effusion.

(1) Infectious Diseases F.J. Muñiz Hospital, Buenos Aires, Argentina.
(2) National Academy of Medicine, Buenos Aires, Argentina.
Correspondence to: Humberto Metta, Tucumán 1630, 3° "C", Postal code C1050AAH, Buenos Aires, Argentina. E-mail: hmetta@sinectis.com.ar

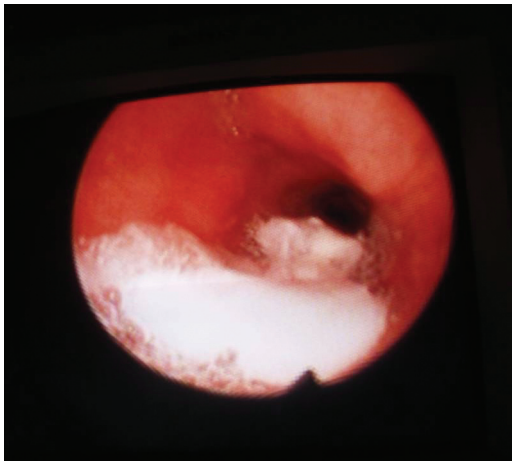

Fig. 2 - Fiberoptic bronchoscopic examination revealed a left endobronchial lobulated tumor that obstructed the ostium of the left upper lobe.

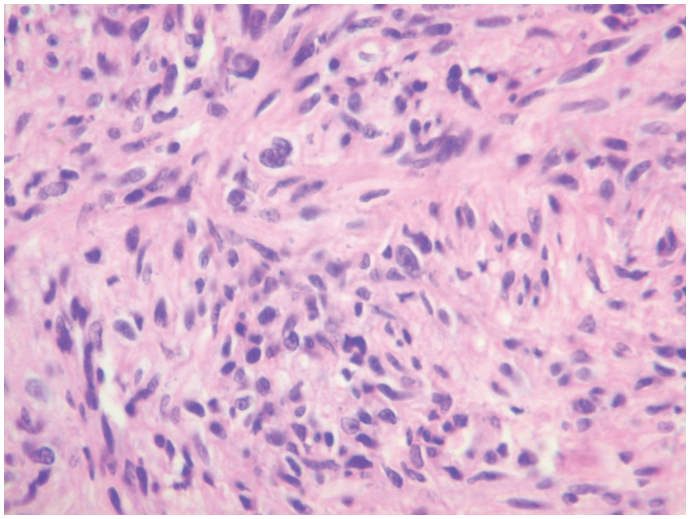

Fig. 3 - A fusocellular proliferation of spindle cells with cigar-shaped nuclei, eosinophilic fibrillar cytoplasms and hialinosis areas, compatible with smooth muscle tumor.

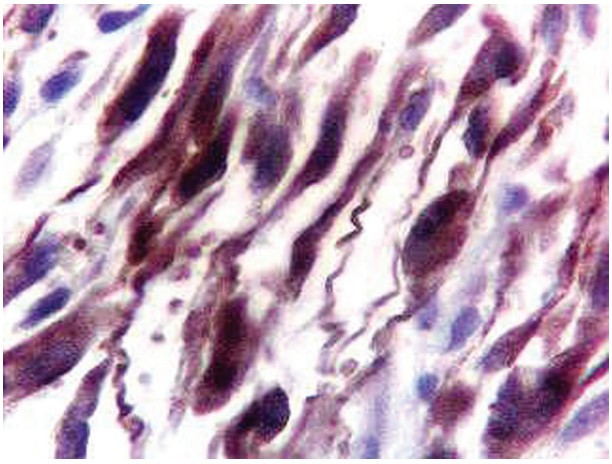

Fig. 4 - The tumor cells showing immunoreactive for specific antismooth muscle actin and HHF35 actin expression.

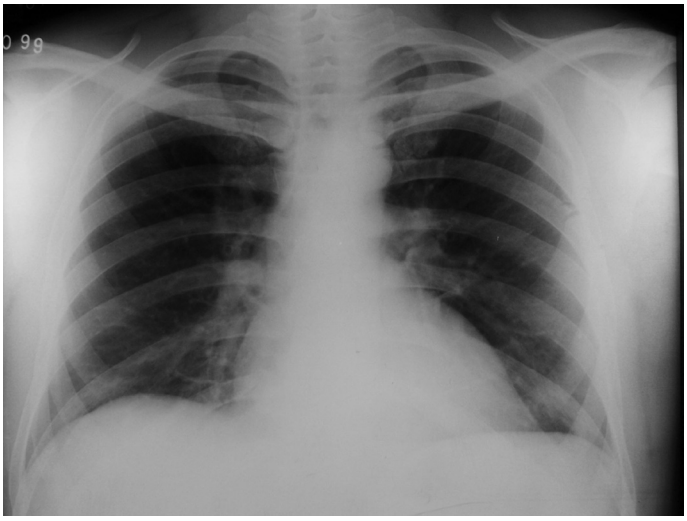

Fig. 5 - Chest radiograph seven months after the tumor eradication.

and hialinosis areas, compatible with smooth muscle tumor (Fig. 3). No mitosis was found in the biopsy specimen. There was relation between the tumor and the smooth muscle with the bronquial wall. The tumor cells showed immunoreactive for specific antismooth muscle actin and HHF35 actin expression (Fig. 4), with no expression of CD117, S-100, CD34 and HMB45, consistent with the histopathological diagnosis of leiomyoma. The detection of EBV by *in situ* hibridization (ISH) for EBV-encoded RNA-1 (EBER-1) and for EBV associated latent membrane protein-1 (LMP-1) by immunohistochemical (IHQ) in formalin fixed, parafin embebed tissue, were negative. During a twelve-month follow-up period, the patient upholds normal respiratory function with no evidence of tumor recurrence (Fig. 5).

DISCUSSION

Tumors of smooth muscle origin rarely involve the lung. Among all tumors surgically excised from the lung, only 4% are benign and, of these, only 2% are leiomyomas, a benign form of smooth muscle tumor[3,15].

Human immunodeficiency virus (HIV)-infected adults have an increased risk to develop some non-AIDS defining neoplasms, as Hodgkin lymphoma and lung carcinoma, but leiomyoma remains rare and generally involves extrapulmonary sites[1,3,6,10,11,12,14,16]. In adults, these tumors should be described in immunocompromised patients with renal transplantation, steroid therapy and AIDS[5].The first case of an endobronchial leiomyoma in an adult with AIDS was described by BLUHM *et al*.[3] in 1997. In the lung, leiomyoma usually involves the bronchi or alveolar wall and it is presented as a solitary lesion, as in the patient we are describing. However, in the case which was presented by BLUHM the neoplasm was multifocal[3].

Endobronchial lesions in adults with HIV include malignancies as Kaposi´s sarcoma and non-Hodgkins lymphoma and several infectious diseases as tuberculosis and other mycobacteriosis[4]. Due to the similar

aspects of bronchoscopic appearance of these different complications of advanced HIV/AIDS disease, the endobronchial or bronchoscopic lung biopsies with special stains (Ziehl-Neelsen, Grocott, Giemsa), should be performed to guarantee the final diagnosis.

Definitive criteria for determining malignancy of SMT, specifically leiomyosarcoma located outside the digestive and genitourinary tracts, includes the size, cellularity, cytologic atypia, necrosis grade and the presence of hemorraghic areas. The most important predictor of malignancy is the level of mitotic activity in the atypical cells that varies by the organ involved[6].

The relationship between EBV infection and the risk of the development of smooth muscle tumors have been well described in the medical literature, especially in tumors affecting children with AIDS and immunosuppressed individuals after liver or renal transplantation[9]. These reports suggest that EBV infection may play a role in the pathogenesis of these tumors in immunocompromised children. Also, the presence of EBV genome in the tumor tissue from HIV-infected adults should be shown and suggests that EBV is implicated in the development of tumors of the smooth muscle in immunocompromised adults[3].

However, there were several reports in which leiomyomas were not related with EBV and ISH was negative, as in our patient[16]. Such features were reported in non-AIDS and immunocompetent hosts[2,8]. These cases marked that other mechanisms also may contribute to develop this kind of tumors, including the retrovirus HIV, in a direct or indirect way[9]. The tumor present in the case we are describing is not identical, in strict sense, to those with EBV associations that are common to AIDS.

In conclusion, endobronchial leiomyoma is still a rare benign tumor in HIV-infected adult patients. Histopathological features are the source to certify the final diagnosis, which is difficult to be suspected only of clinical expression.

Our report suggests that, if the diagnosis of leiomyoma is more an exception than a rule, SMT should be included in the differential diagnosis of endobronchial masses associated with atelectasis in HIV/AIDS patients.

RESUMO

Leiomioma endobronquial: rara neoplasia não definida em paciente com AIDS

Neoplasmas da musculatura lisa são mais freqüentes em crianças infectadas pelo vírus da imunodeficiência humana do que em adultos HIV-soropositivos. Leiomioma endobronquial é um tumor benigno em pacientes adultos infectados por HIV. Vírus Epstein-Barr (EBV) tem sido implicado na patogenia destes tumores. Descrevemos paciente adulto infectado pelo HIV com atelectasia do lobo pulmonar superior esquerdo como primeira manifestação clínica de leiomioma intrabronquial. Neste caso não pudemos demonstrar a associação com EBV. Nosso relato sugere que tumores de musculatura lisa como leiomioma deveriam ser incluídos no diagnóstico diferencial de massas endobronquiais em pacientes com AIDS.

REFERENCES

1. BARBASHINA, V.; HELLER, D.S.; HAMEED, M. *et al*. - Splenic smooth-muscle tumors in children with acquired immunodeficiency syndrome: report of two cases of this unusual location with evidence of an association with Epstein-Barr virus. **Virchows Arch., 436:** 138-139, 2000.

2. BILGIN, S.; YILMAZ, A.; OKUR, E. *et al.* - Primary endobronchial leiomyoma: a case report. **Tüberk. Toraks., 52**: 272-274, 2004.

3. BLUHM, J.M.; YI, E.S.; DIAZ G.; COLBY, T.V. & COLT, H.G. - Multicentric endobronchial smooth muscle tumors associated with the Epstein-Barr virus in an adult patient with the acquired immunodeficiency syndrome. **Cancer, 80:** 1910-1913, 1997.

4. CORTI, M.E.; VILLAFAÑE, M.F.; TRIONE, N.; SCHTIRBU, R. & NARBAITZ, M. - Primary pulmonary AIDS-related lymphoma. **Rev. Inst. Med. trop. S. Paulo, 47:** 231-234, 2005.

5. DEYRUP, A.T.; LEE, V.K.; HILL, C.E. *et al*. - Epstein-Barr virus associated smooth muscle tumors are distinctive mesenchymal tumors reflecting multiple infection events: a clinicopathologic and molecular analysis of 29 tumors from 19 patients. **Amer. J. surg. Path., 30:** 75-82, 2006.

6. ENZINGER, F.M. & WEISS, S.W. - Benign tumors of smooth muscle. In: ENZINGER, F.M. & WEISS, S.W., ed. **Soft tissue tumors**. 3. ed. St. Louis, C.V. Mosby, 1995. p. 467-490.

7. JENSON, H.B.; LEACH, C.T.; McCLAIN, K.L. *et al*. - Benign and malignant smooth muscle tumors containing Epstein-Barr virus in children with AIDS. **Leuk. Lymphoma, 27:** 303-314, 1997.

8. KO, S.M.; HAN, S.B.; LEE, S.K. *et al.* - Calcified endobronchial leiomyoma. **Brit. J. Radiol., 80**: e91-93, 2007.

9. LEE, E.S.; LOCKER, J.; NALESNIK, M. *et al*. - The association of Epstein-Barr virus with smooth-muscle tumors occurring after organ transplantation. **New Engl. J. Med., 332:** 19-25, 1995.

10. MORGAN, B.K.; COMPTON, C.; TALBERT, M.; GALLAGHER, W.J. & WOOD, W.C. - Benign smooth muscle tumors of the gastrointestinal tract: a 24-year experience. **Ann. Surg., 211:** 63-66, 1990.

11. STARASOLER, L.; VUITCH, F. & ALBORES-SAAVEDRA, J. - Intranodal leiomyoma: another distinctive primary spindle cell neoplasm of lymph node. **Amer. J. clin. Path., 95:** 858-862, 1991.

12. STEEL, T.R.; PELL, M.F.; TURNER, J.J. & LIM, G.H.K. - Spinal epidural leiomyoma occurring in an HIV-infected man: a case report. **J. Neurosurg., 79:** 442-445, 1993.

13. VOGEL, J.; HINRICHS, S.H.; REYNOLDS, B.K.; LUCIW, P.A. & JAY, G. - The HIV tat gene induces dermal lesions resembling Kaposi's sarcoma in transgenic mice. **Nature, 335:** 606-611, 1988.

14. WACHSBERG, R.H.; CHO, K.C. & ADEKOSAN, A. - Two leiomyomas of the liver in an adult with AIDS: CT and MR appearance. **J. comput. Assist. Tomogr., 18:** 156-167, 1994.

15. WHITE, S.H.; IBRAHIM, N.B.N.; FORRESTER-WOOD, C.P. & JEYASINGHAM, K. - Leiomyomas of the lower respiratory tract. **Thorax., 40:** 306-311, 1985.

16. ZETLER, P.J.; FILIPENKO, J.D.; BILVEB, J.H. & SCHMIDT, N. - Primary adrenal leiomyosarcoma in a man with acquired immunodeficiency syndrome (AIDS). Further evidence for an increase in smooth muscle tumors related to Epstein-Barr infection in AIDS. **Arch. Path. Lab. Med., 119:** 1164-1167, 1995.

Received: 23 January 2008
Accepted: 30 October 2008